SYSTEMIC AND PULMONARY VENOUS CONNECTIONS IN VISCERAL HETEROTAXY WITH ASPLENIA

Diagnostic and surgical considerations based on seventy-two autopsied cases

To facilitate the preoperative diagnosis and surgical management of visceral heterotaxy and asplenia, 72 postmortem cases were reviewed with particular attention focused on the systemic and pulmonary venous connections. The superior vena cava was bilateral in 51 cases (71%), but in 9 cases one of the superior venae cavae was partly or totally atretic. Patent bilateral superior venae cavae were found in 42 cases (58%) and the superior vena cava was unilateral in 21 (29%). Although the inferior vena cava was never interrupted, a prominent azygos vein was found in 6 cases (8%). Some hepatic veins drained separately from the inferior vena cava in 20 cases (28%). An intact coronary sinus was rare (2 cases, 3%). Anomalous pulmonary venous connection to a systemic vein was total in 42 (58%) of 72 and partial in 2 (3%) of 72, with obstruction in 24 (55%) of 44. Abnormal pulmonary artery branches (severe hypoplasia, localized stenosis, or discontinuity) were present in 21 (29%), and these obstructive arterial anomalies were associated with a significantly higher prevalence of anomalous pulmonary venous connection ($p < 0.01$) and of pulmonary venous obstruction ($p < 0.01$). Cardiac pulmonary venous connections were found in 28 (39%), with the pulmonary veins and the inferior vena cava entering the same atrium in 10 (36%) of 28. (J THORAC CARDIOVASC SURG 1995;110:641-50)

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Visceral heterotaxy with congenital absence of the spleen and complex cardiac malformations (asplenia syndrome) is a relatively rare condition that occurs in 2.8% of the cases with congenital heart disease in the Cardiac Registry of the Children's Hospital in Boston. The prognosis of these patients is extremely poor with a 79% mortality rate during the first year of life.[1]

Although the usual patterns of the systemic and pulmonary venous connections, the atrial and ventricular morphologic features, and the atrioventricular and ventriculoarterial alignments have been described in numerous publications,[1-10] there is little detailed information concerning the wide variation that exists in the systemic and pulmonary venous connections, which is a matter of considerable surgical importance in patients with asplenia syndrome.

Our report focuses on a detailed description of the wide variety of the systemic and pulmonary venous connections, which has practical relevance because the main goal of the various surgical procedures is to separate the systemic and pulmonary circulations.[5, 11-21] We also studied the correlation between the pulmonary arterial patterns and the presence or absence of anomalous pulmonary venous connections with or without pulmonary venous obstruction.

Material and methods

Among the records of the 3020 cases of congenital heart disease in the Cardiac Registry of the Children's Hospital of Boston, 86 were reported as having congenital asplenia (2.8%).* The present report is based on 72 of these 86 cases; 78 were available for reexamination, but 6 were excluded because poor preservation made accurate assessment of the systemic and pulmonary venous connections impossible.

All 72 heart specimens were examined with particular emphasis on the systemic and pulmonary venous connec-

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*Sixty of these cases were previously reported, two of them individually[22, 23] and 58 as part of a study of all types of visceral heterotaxy.[2, 4]

tions. A brief description of the atrial appendages and atrial septa is also given. The diagnosis of the anatomic identity of the atria, that is, the atrial situs, has been the subject of previous publications[2, 4] and is not included in this report.

We reviewed the postmortem reports and clinical summaries to obtain information regarding sex, age at death, operation, and pulmonary venous obstruction. The statistical analysis for the data presented in Tables I and II was done with the χ^2 test with the Yates correction or Fisher's exact test. A p value less than 0.05 was considered statistically significant.

Results

The male/female ratio was 1.8:1.

The median age at death was 35 days and ranged from 5 hours to 35⁹⁄₁₂ years; 35% of patients died in the first week of life. When pulmonary outflow tract atresia or pulmonary venous obstruction was present, the median ages at death were 4 days and 7 days, respectively. In contrast, when pulmonary outflow tract atresia and pulmonary venous obstruction were not present, the median ages at death were 2.6 months and 3 months, respectively.

Forty-two patients underwent cardiac operation with a total of 61 operations. There was considerable difference in the median age at death of patients who underwent operation (5 months) and that of those who did not (5 days). The median age at the first operation was 28 days. Nevertheless, 40% of patients required operation during the first week of life.

Systemic veins. The various types of systemic venous connections are diagrammatically presented in Fig. 1.

A normal coronary sinus (CoS) is rare in visceral heterotaxy with asplenia and was present in only two cases of this series (2 [3%] of 72). These two cases exemplify the linkage we have previously reported between the CoS and the atrioventricular canal,[2] that is, all cases of visceral heterotaxy and asplenia with an unroofed CoS have a common atrioventricular canal and a common atrioventricular valve. All cases with a normal CoS have two separate and usually normal atrioventricular valves. We present the visceral and cardiac malformations of these two rare cases in Table I.

Bilateral superior venae cavae (SVCs) were present in 51 (71%) of 72 specimens. In 9 cases one of the two SVCs was atretic. When bilaterally patent (42 cases, 58%), both SVCs opened directly into the roof of the atria immediately posterior to the atrial appendages. Exceptions were the two cases with a normal CoS. In these two cases one SVC drained directly into the anatomically right atrium and the other indirectly via the normal CoS.

The sizes of the two SVCs showed great variation (Fig. 1). Both SVCs were well developed in 28 (39%) of 72 cases (columns 1 and 2, Fig. 1). In 14 (19%) of 72, one of the two SVCs was moderately or markedly underdeveloped (columns 3 and 4, Fig. 1). In 9 (12.5%) of 72, one of the two SVCs was atretic (column 5, Fig. 1). In the remaining 21 (29%) of 72, only one SVC was identified (column 6, Fig. 1). Hence, unequal SVCs or a single patent SVC was present in 44 (61%) of 72 cases.

The inferior vena cava (IVC) was more often ipsilateral with the larger of the two SVCs, or with the single patent SVC, as in 27 (61%) of 44 cases (rows A, C, E, F, and G, Fig. 1). The IVC was contralateral to the single patent SVC or to the larger of the two SVCs in 17 (39%) of 44 cases (rows B, D, and H, Fig. 1). When one of the two SVCs was very small (column 4, Fig. 1), an innominate vein was present in all but one case.

The IVC was always uninterrupted. Nevertheless, in six cases (8%) a prominent azygos vein connected with the left SVC (3 cases), with the right SVC (2 cases), or with both SVCs (1 case).

Some of the hepatic veins drained separately from the IVC into the contralateral atrium in 18 (25%) of 72 cases or into the same atrium in 2 (3%) of 72 cases (rows E, F, G, and H, Fig. 1). The IVC and the hepatic veins interconnected via a large venous sinus just before their separate entry into the atria in four cases. This assessment could not be made in all the cases because of postmortem artifacts.

Atria. The size and shape of the atrial appendages was never completely normal. In 51 cases (71%) both appendages had a similar shape, but not always a similar size, and they usually resembled the right atrial appendage. In the remaining 21 specimens (29%), the shape of the appendages was dissimilar and resembled solitus or inversus atrial arrangement. The size and position of the appendages relative to each other could be assessed in 60 cases. We found that the larger and more anterior appendage was right sided in 38 (63%) of 60 specimens and left sided in 22 (37%) of 60. The IVC drained into the atrium with the larger and more anterior appendage in 37 (62%) of 60 cases. In the remaining 12 (17%) of 72 cases the size and position of the atrial appendages were either similar or could not be assessed because of surgical or postmortem artifacts.

The atrial septum was extremely underdeveloped (39 cases) or completely absent (8 cases) in 47

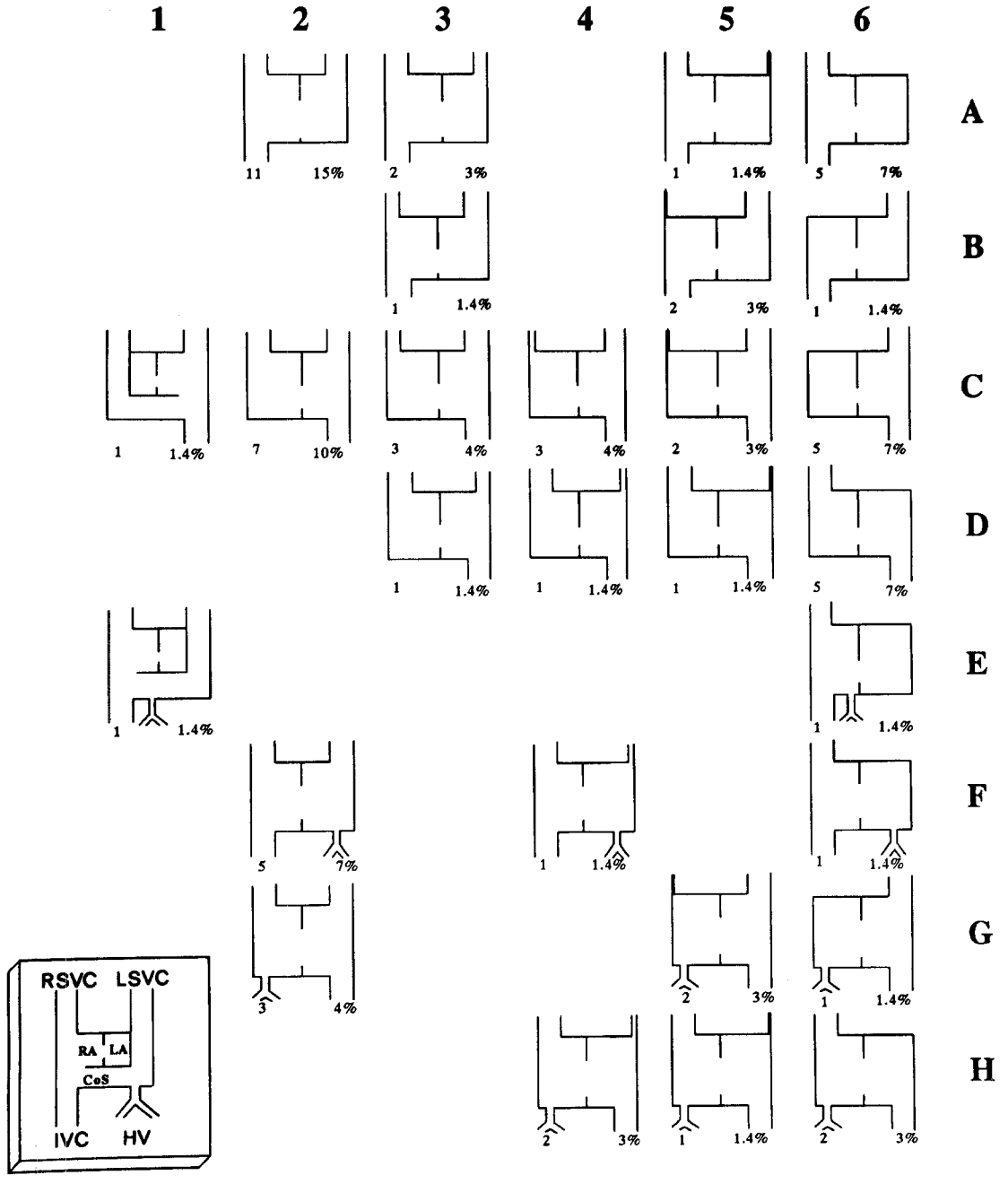

Fig. 1. Diagrammatic presentation of systemic venous connections in 72 cases of visceral heterotaxy with asplenia. Normal CoS was present in two cases *(column 1). Vertical columns* outline difference in size between two SVCs (equal: *columns 1* and *2;* unequal: *columns 3* and *4;* one atretic: *column 5;* single: *column 6*). *Horizontal rows* outline absence *(rows A, B, C, D)* or presence *(rows E, F, G, H)* of separate drainage of the hepatic veins and the relationship between IVC and larger of two SVCs (ipsilateral: *rows A, C, E, F, G;* contralateral: *rows B, D, H*). *HV,* hepatic veins; *LA,* morphologically left atrium; *LSVC,* left SVC; *RA,* morphologically right atrium; *RSVC,* right SVC.

(65%) of 72 specimens. When extremely underdeveloped, it was usually represented by a single strand of fibromuscular tissue extending from the orifice of the IVC to the anterior atrial wall at the level of the common atrioventricular valve anulus or just medially to the orifice of the SVC. Atrial septal components were present in 25 (35%) of 72 cases. An ostium primum defect, with or without an ostium

Table I. *Visceral and cardiac findings in asplenia with normal CoS*

	Case 1: 7-month-old male infant with dextrocardia	*Case 2: 40-hour-old female infant with levocardia*
Abdominal viscera	Symmetric liver	Solitus liver
	Left-sided gallbladder	Heterotopic pancreas
	Centrally located pancreas	Cervical and vaginal atresia
	Right-sided stomach and rectum	Bicornuate uterus
	Common mesentery	Asplenia
	Asplenia	
Lung lobation	Inversus	Bilaterally bilobed
Bronchi	Inversus	Tracheal agenesis
		Bronchoesophageal fistula connects lower end of esophagus with 2 bronchi of equal length*
Atrial appendages	JAA (rt)	Atypical shape
	RAA superior to LAA	RAA much larger than left
	Atypical shape	RAA more anterior than LAA
	Equal size	Crista terminalis with RAA
	Crista terminalis unidentifiable†	
Systemic veins	CoS enlarged, receives persistent RSVC	Bilateral azygos veins connect with ipsilateral SVCs
	LIVC, LSVC, and CoS connect with RA (left)	RIVC, RSVC, and CoS and LSVC connect with RA (left)
Pulmonary veins	Poor incorporation of pulmonary venous chamber into LA (right) via a single orifice	Normally connected with LA
Atrial situs	Inversus	Solitus
Atrial septum	Small remnant of septum primum attaches on the ridge between the two appendages	Patent foramen ovale
	Large ostium secundum defect	
AV valves	Normal	Normal
Ventricles	D-loop (AV situs concordance with AV alignment discordance)	D-loop (AV concordance)
	Superoinferior ventricles	RVH
	RVH, RV inflow underdeveloped	CS malalignment VSD
	Very large inlet and CS malalignment VSD	Sub-PS
	LVH	
	Sub-PS	
Semilunar valves	PV bicuspid, stenotic	PV bicuspid, stenotic
	Ao valve normal	Ao valve normal
Great arteries	Right Ao arch	Left Ao arch
	Single orifice for the two common carotids	Aberrant right SCV
	Aberrant left SCV	
VA relationship	DORV	DORV

Ao, Aortic; *AV,* atrioventricular; *CS,* conal septum; *D,* dextro; *DORV,* double outlet right ventricle; *JAA,* juxtaposition of the atrial appendages; *L,* levo; *LA,* left atrium; *LAA,* left atrial appendage; *LIVC,* left IVC; *LSVC,* left SVC; *LVH,* LV hypertrophy; *PS,* pulmonary stenosis; *PV,* pulmonary valve; *RA,* right atrium; *RAA,* right atrial appendage; *RIVC,* right IVC; *RSVC,* right SVC; *RV,* right ventricle; *RVH,* right ventricular hypertrophy; *SCV,* subclavian; *VSD,* ventricular septal defect.

*The abnormal origin of the bronchi from the lower part of the esophagus eliminated any bronchoarterial relationship.

†The crista terminalis represents the junctional muscular crest between the atrial appendages and the adjacent SVC. In this case the right atrial appendage was right sided and the right atrial SVC was left sided. Hence a crest between the two was not possible. The persistent right SVC continued into the normal CoS and thus could not form a terminal crest with either appendage. This case supports the view that the crista terminalis is not always the essential landmark for the anatomic right atrium.

secundum defect, was present in all but the two cases with a normal CoS. One of these two cases had a patent foramen ovale and the other had an ostium secundum defect (Table I).

Pulmonary veins. The pulmonary venous connections showed great variation. We present them in three groups according to whether they had cardiac, mixed, or extracardiac connections (groups A, B, and C in Figs. 2 and 3).

Group A. Group A, which had only cardiac (atrial) connections, included 28 (39%) of 72 cases (cases 1 through 28, Fig. 2).

The pulmonary veins connected normally with four orifices in the back of the atrium in 12 (43%) of

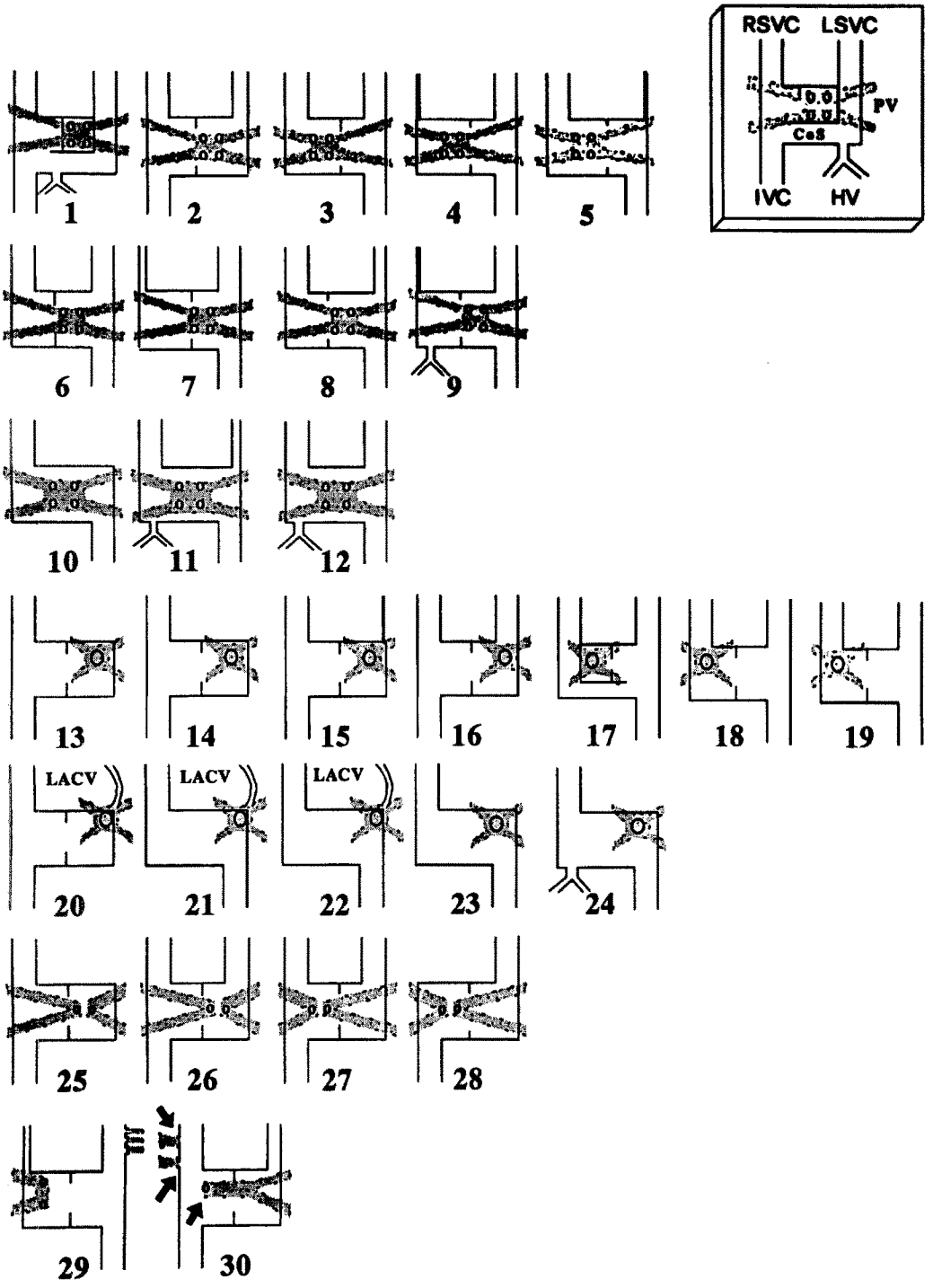

Fig. 2. Diagrammatic presentation of systemic and pulmonary venous connections in 30 cases of visceral heterotaxy with asplenia associated with total or partial cardiac pulmonary venous connections (groups A and B). In cases 1 through 12 pulmonary veins connect with heart normally via four orifices in posterior atrial wall. In cases 13 through 28 pulmonary veins connect with atrial segment of heart via single or double orifice (incomplete incorporation of common pulmonary vein). In cases 29 and 30 pulmonary veins connect partly with atria and partly with SVC. *Arrows* indicate sites of obstruction of pulmonary venous drainage. *PV,* Pulmonary veins; *LACV,* levoatrial cardinal vein. Other abbreviations as in Fig. 1.

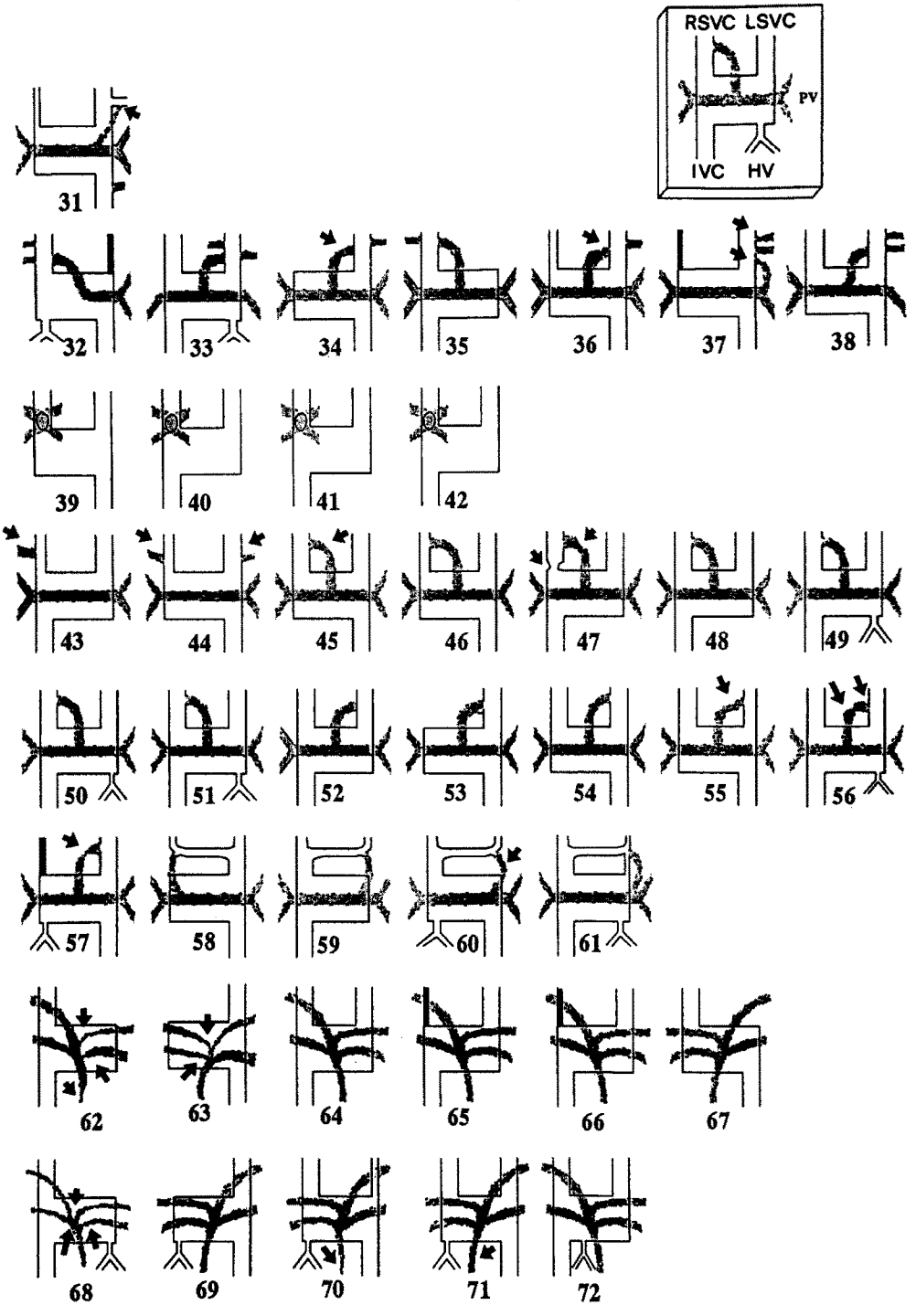

Fig. 3. Diagrammatic presentation of systemic and pulmonary venous connections in 42 cases of visceral heterotaxy with asplenia associated with extracardiac pulmonary venous connections (group C). In case 31 abnormal pulmonary venous connections are mixed, partly to azygos vein and partly to IVC. In cases 32 through 61 abnormal pulmonary venous connections are supracardiac and in cases 62 through 72 they are subdiaphragmatic. *Arrows* show sites of obstruction of pulmonary venous drainage. Abbreviations as in Fig. 1.

28 (cases 1 through 12, Fig. 2). In four cases (cases 6 through 9, Fig. 2) this connection was with the atrium that also received the IVC. In five cases (cases 1 through 5, Fig. 2) the pulmonary veins entered the atrium contralateral to the IVC entry, and in the remaining three cases (cases 10 through 12, Fig. 2) surgical or postmortem artifacts made impossible the assessment of their relationship with the atrial septum.

In 16 (57%) of 28 cases the pulmonary veins connected with the atrial part of the heart via a single (cases 13 through 24, Fig. 2) or double (cases 25 through 28, Fig. 2) orifice, that is, poor incorporation of the common pulmonary vein. In these 16 cases the veins from both lungs connected behind the atria to form a common chamber, which drained into one of the two atria by a single, or double, generally unrestrictive orifice (cases 13 through 28, Fig. 2). As a rule, the site of the connection was abnormally high, on the roof of the atrium, near the atrial appendage, or close to the orifice of the SVC. In six cases the connections were with the atrium that also received the IVC and in 10 cases with the atrium that was contralateral to the IVC entry. A levoatrial cardinal vein[24] connecting the pulmonary venous confluence with the ipsilateral innominate vein was present in three of the cases with single orifice connection (cases 20 through 22, Fig. 2).

Group B. Group B, which had cardiac and extracardiac pulmonary venous connections, included only two cases (cases 29 and 30, Fig. 2). In both, the one lung drained into the atrial part of the heart and the other into the ipsilateral or contralateral SVC via multiple orifices, which were restrictive in one case (case 30, Fig. 2).

Group C. Group C was the largest and included 42 (58%) of 72 cases. The connection of the pulmonary veins to a systemic vein was supracardiac in 30 (71%) of 42 and infradiaphragmatic in 11 (26%) of 42 cases. In a single case (case 31, Fig. 3), the left lower lobe drained into the left-sided IVC and all the other pulmonary veins formed a confluence that connected with the left azygos vein via a single and restrictive orifice.

There was great variation in the site and mode of connection of the pulmonary veins with the systemic veins and they are individually outlined in Fig. 3.

The pulmonary venous drainage was into the right SVC in 14 cases, left SVC in 11, portal vein in 4, ductus venosus in 4, left innominate vein in 3, gastric vein in 2, right innominate vein in 1, both SVCs in 1, and an unknown abdominal vein in 1. As a rule, all the pulmonary veins formed a venous confluence behind the atria, which in turn connected with a systemic vein via a single vertical or oblique vein. Nevertheless there were seven cases in which one lung or part of one lung drained into the systemic vein via a separate pathway (cases 32 through 38, Fig. 3). In 4 of 42 cases the pulmonary venous confluence was formed at the level of the SVC-atrial junction and connected with the SVC via a single unrestricted orifice (cases 39 through 42, Fig. 3). In two instances (cases 43 and 44, Fig. 3) there was no direct connection between the pulmonary venous confluence and the atria or any of the systemic veins. The pulmonary venous drainage appeared to be accomplished via a small vein from the right upper lobe to the right SVC in one case and from the upper lobe of each lung to the ipsilateral SVC in the other. The smallness of this vein constituted the site of pulmonary venous obstruction.

Clinical or anatomic findings of pulmonary venous obstruction were also present in all 11 subdiaphragmatic connections (cases 62 through 72, Fig. 3) and in 10 of the supracardiac connections (cases 31, 34, 36, 37, 45, 47, 55 through 57, and 60; Fig. 3). Compression of the vertical or oblique connecting vein between the bronchi and the pulmonary artery branches (6 cases), compression between the bronchi and the aortic arch (1 case), or stenosis of the orifice of the vertical vein (3 cases) was the cause of pulmonary venous obstruction in the supracardiac group. The pathway through the liver was the constant site of obstruction in the group with subdiaphragmatic connections and a closed ductus venosus. In addition, the connection of the vertical vein with the ductus venosus, the portal vein, or the gastric vein was as a rule restrictive. In three cases (cases 62, 63, and 68; Fig. 3) there was additional stenosis of the individual pulmonary veins (indicated by arrows, Fig. 3).

All the cases with pulmonary venous obstruction were in groups B and C, that is, the groups with partial or total extracardiac pulmonary venous connections (Figs. 2 and 3). The prevalence was 24 of 44, or 55%.

The relationships between the patterns of pulmonary arterial anatomy and the presence or absence of pulmonary outflow obstruction are presented in Table II. The prevalence of anomalous pulmonary venous connections to systemic veins and that of pulmonary venous obstruction in relation to the morphologic features of the pulmonary arteries are presented in Table III. When the pulmonary artery

Table II. *Incidence of normal and abnormal pulmonary artery branches in 72 postmortem cases of asplenia in relation to the presence or absence of pulmonary outflow obstruction*

	Normal PA branches		*Abnormal PA branches*	
Pulmonary outflow	*n*	*%*	*n*	*%*
No obstruction ($n = 5$*)	5/5	100	0	0
Pulmonary stenosis ($n = 41$)	33/41	80	8/41	20
Pulmonary atresia ($n = 26$)	13/26	50	13/26	50
Total ($n = 72$)	51/72	71	21/72	29

Normal pulmonary artery branches are pulmonary artery branches of normal size or slightly hypoplastic, but without localized stenosis or discontinuity. Abnormal pulmonary artery branches are pulmonary artery branches with localized stenosis or discontinuity, or both. *PA,* Pulmonary artery.

*Three of these cases had aortic atresia and large pulmonary arteries.[22, 23] The other two had double-outlet right ventricle without aortic or pulmonary outflow obstruction.

Table III. *Prevalence of anomalous pulmonary venous connection and pulmonary venous connection obstruction in 72 postmortem cases of asplenia in relation to the morphology of the pulmonary artery branches*

	APVC		*APVC with obstruction*	
Morphology of PA branches	*n*	*%*	*n*	*%*
Normal PAs ($n = 51$)	26/51	51	12/51	24
Abnormal PAs ($n = 21$)	18/21	86	12/21	57
Total ($n = 72$)	44*/72	61	24/72	33

The differences in the prevalences of anomalous pulmonary venous connection and of pulmonary venous obstruction between normal and abnormal pulmonary artery branches are statistically significant; $p < 0.01$. *PA,* Pulmonary artery; *APVC,* anomalous pulmonary venous connections. Other explanations as in Table II.

*Two cases had partial anomalous pulmonary venous connection to a systemic vein and 42 had total anomalous pulmonary venous connection to systemic veins.

branches did not have localized obstruction or discontinuity, the prevalence of anomalous pulmonary venous connection was 51%. However, when the pulmonary artery branches showed abnormalities such as severe hypoplasia, localized stenosis, or discontinuity, the prevalence of anomalous pulmonary venous connection was 86% (Table III). This difference was statistically significant ($p < 0.01$). Similarly when the pulmonary artery branches were normally formed (that is, without localized stenosis or discontinuity), the prevalence of pulmonary venous obstruction was 24% (Table III). When the pulmonary artery branches were abnormal, the prevalence of pulmonary venous obstruction was 57% (Table III). This difference was also statistically significant ($p < 0.01$).

Discussion

Specific diagnostic information concerning the systemic and pulmonary venous connections and the pulmonary arterial pattern constitutes the essential background for planning any surgical treatment of patients with visceral heterotaxy and asplenia.[5, 17-21, 25]

In this and other studies[2, 4, 26] the pulmonary venous connections were considered abnormal when the pulmonary veins maintained their early embryonic connections with the systemic veins.

When the pulmonary veins connect with the atrial segment of the heart, one has reason to conclude that the common pulmonary vein has developed; hence the connection of the pulmonary veins with the heart is normal.[2, 4, 26]

The normalcy of the pulmonary venous connections in the cases of common atrium or the cases of pulmonary veins draining into the right atrium is supported by the following anatomic findings. Normally or abnormally draining pulmonary veins appear to connect with the same area of the posterior atrial wall. This area is located between the two SVCs, when bilateral SVCs are present, to the left of a single right SVC, or to the right of a single left SVC.

Malposition of the septum primum toward the anatomically left atrium will allow half or all of the pulmonary veins to drain into the right atrium even though they are connected normally with what otherwise would have been the posterior wall of the left atrium.[26, 27] Absence or marked underdevelopment of the septum secundum, which usually receives the normal attachments of the septum primum, appears to be the reason for the malposition of septum primum.[27]

When the atrial septa (primum and secundum) are absent, as in many cases in this study, the pulmonary veins enter the back of the common atrium in the area between the two SVCs, to the left of a single right SVC, or to the right of a single left SVC. Hence the pulmonary veins are normally connected to the posterior atrial wall. If the atrial

septa had developed, this part of the atrial wall would belong to the left atrium.

When the pulmonary veins connect with the atrial segment of the heart via one or two orifices then the incorporation of the common pulmonary vein into the left atrium is incomplete but not abnormal.

Our study indicates that the two malformations that constitute the most severe problems in the newborn period, namely pulmonary artery stenosis or atresia and anomalous pulmonary venous connections with or without obstruction, tend to occur together. The abnormal morphology of the pulmonary artery branches showed a statistically significant positive correlation with the occurrence of anomalous pulmonary venous connections and with the occurrence of pulmonary venous obstruction. Specifically, when the pulmonary arteries were markedly hypoplastic, stenotic, or discontinuous, an anomalous pulmonary venous connection was more probable than when the pulmonary arteries were slightly hypoplastic or normal (86% versus 51%, respectively; $p < 0.01$, Table III). Also, obstruction of the anomalous pulmonary venous connection was more probable when the pulmonary arteries were abnormal than when the pulmonary arteries did not have localized obstructions or discontinuity (57% versus 24%, respectively; $p < 0.01$, Table III).

These linkages should be taken into consideration when the possibilities of abnormal pulmonary venous connections and pulmonary venous obstruction are investigated.

When the pulmonary venous confluence connected with a systemic vein additional pulmonary venous connections to the SVC or IVC were present in several cases (cases 31 through 38, Fig. 3). The diagnosis of these additional pulmonary venous connections is essential to plan the appropriate surgical procedure.

Many authors have reported that a bidirectional cavopulmonary shunt placed at a young age is helpful in preparation for the Fontan operation.[28-30] When such a procedure is contemplated in patients with visceral heterotaxy and asplenia, a preoperative diagnosis of an enlarged azygos vein with an intact IVC (a situation that occurred in 6 cases of this series) should be differentiated from that of an enlarged azygos and interrupted IVC to decide whether the azygos vein should be ligated.

The gratifying early results obtained with the bidirectional cavopulmonary shunt operation in cases with interrupted IVC[31] have inspired some surgeons to perform a subtotal cavopulmonary connection with partial or total hepatic vein exclusion.[32, 33] Many patients with asplenia could be ideal candidates for this option because of the separate drainage of some or all of the hepatic veins and the IVC (28% in this series). If such a procedure is planned, one should investigate the presence or absence of an interconnecting venous sinus between the IVC and the hepatic veins just below their separate entry to the heart. Biventricular repair or univentricular physiologic repair of cases with visceral heterotaxy and asplenia have been done in recent years.[11, 13, 15, 17-21] It is hoped that the great variations of the systemic and pulmonary venous connections outlined in this report will stimulate their careful and detailed preoperative assessment in patients with visceral heterotaxy and asplenia. This assessment is essential in deciding the operability of the case and the most appropriate operative procedure.

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